

A Pediatric Case of Neuromyelitis Optica and Pulmonary Inflammatory Myofibroblastic Tumor

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Conflict of Interest

The authors have no conflicts of interest to disclose.

Consent for Publication

The parents gave consent for this case report to be written up and published.

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To the editor,

Neuromyelitis optica spectrum disorders (NMOSD) is a rare autoimmune demyelinating disorder primarily affecting the spinal cord and optic nerve. Pediatric patients account for 4% of all NMOSD cases, with the average age of onset at 12 years old¹. On the other hand, Inflammatory myofibroblastic tumor (IMT) is an uncommon, slow-growing neoplasm, comprising around 0.1% of all neoplasms³. IMT typically impacts the lungs in children and young adults⁵. Despite documented cases of pulmonary manifestations in NMOSD and nervous system involvement in pediatric IMT cases³, the simultaneous occurrence of NMOSD and IMT has not been recorded. In this report, we present a case of NMOSD in a 10-year-old child from central Texas, who was also subsequently diagnosed with an inflammatory myofibroblastic tumor in the left lower lung lobe.

A 10-year-old girl with a medical history of myopia corrected with lenses, visited the emergency department due to a 2–3-day history of left eye blurriness. Initially manifesting as a "line near the bottom of her eye," her vision problems progressed to a larger spot, eventually causing loss of vision in the lower midline quadrant of her left eye. She also experienced vomiting and finger clubbing. Evaluation in the emergency department revealed bilateral papilledema and an afferent pupillary defect in the left eye. Upon further assessment, MRI of the brain, orbits and spine showed left-sided optic neuritis and signal abnormalities in the cervical and thoracic spine. Our patient subsequently had an autoimmune work up, and serum aquaporin-4 testing was the only antibody that returned positive at a titer of 1:100 leading to a diagnosis of NMOSD.

The patient underwent a high-dose methylprednisolone treatment for five days. Given the inadequate improvement in her left eye's vision, plasmapheresis was initiated on hospital day 5. Mild improvement was observed after the third plasmapheresis session on day 8. After her fourth plasmapheresis session, the patient reported her vision in her left eye improved to where she could see outlines but not colors. Due to hypotension with her fourth and fifth plasmapheresis sessions, after her fifth session (hospital day 12), the decision was made to discontinue further plasmapheresis sessions. An intravenous immunoglobulin (IVIG) regimen, 1 g/kg per day, was initiated for two days. By hospital day 15, the patient's left eye could clearly see nearby objects and distinguish outlines from a distance. Following discharge, the patient started rituximab therapy.

One month later, during an outpatient workup by pediatric pulmonology for bilateral finger clubbing, a chest x-ray demonstrated a left lower lobe lung mass. A contrasted CT scan of the chest was performed for further evaluation. It demonstrated a 2.7 x 3.1 x 4.1 cm mass in the left lower lung lobe and fullness of the pancreatic head.

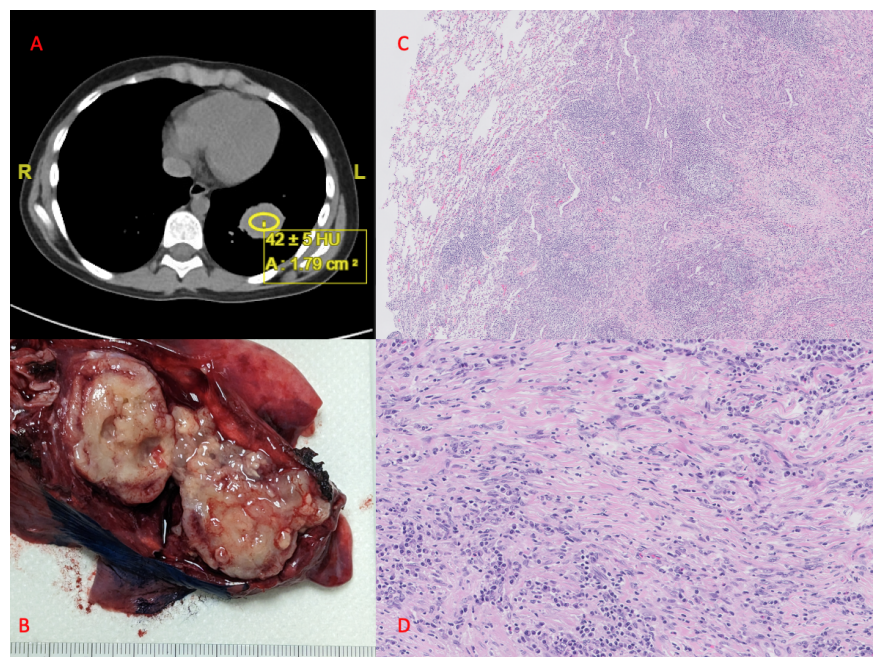


Figure 1: a) initial outpatient CT scan of the chest highlighting the lung mass; b) gross specimen of the lung mass; c) histological image showing spindle cell proliferation with prominent inflammatory infiltrate encroaching on adjacent alveoli; d) histological image highlighting the mixed inflammatory cells including plasma cells, lymphocytes, and eosinophils.

The patient was referred to pediatric surgery and pediatric oncology and she was reviewed at tumor board. A video assisted thoracoscopy (VATS) with left lower lobe lobectomy was performed three months after the initial hospitalization. The left lower lobe (inclusive of the tumor) and associated axillary nodes were removed en bloc in a via thoracotomy. The tumor was sent to pathology without fixation. Postoperatively the patient recovered quickly, with removal of her chest tube and discharge home on the first postoperative day. A post-op chest x-ray demonstrated pneumothorax. She underwent chest tube placement four weeks after her initial operation due to worsening of pneumothorax following discharge from the hospital and had complete, persistent resolution of the pneumothorax.

Pathological examination showed a spindle cell lesion with associated lymphoplasmacytic and histiocytic inflammation. The final pathologic diagnosis made by an outside facility was an inflammatory myofibroblastic tumor (IMT)/inflammatory pseudotumor with microscopically negative margins and negative lymph nodes. The patient still has residual visual abnormalities in the left eye and regularly has imaging to monitor tumor recurrence in the chest and abdomen.

Neuromyelitis optic spectrum disorders and IMT are uncommon conditions, afflicting both adults and children. NMOSD is an CNS autoimmune disorder in which antibodies to the aquaporin-4 water channel are identified in one-half to two-thirds of children¹. IMT are usually benign pulmonary and abdominal neoplasms that most commonly affect children and young adults. While the exact tumor pathogenesis remains uncertain, an overlap exists between IMTs and IgG-4 related disorders. NMOSD is known to be associated with other autoimmune disorders such as systemic lupus erythematosus, Sjogren syndrome, autoimmune thyroid disease, and myasthenia gravis⁴. However, no link between NMO and IMTs has been identified.

NMOSD often presents with visual deficits, sensory dysfunction and constitutional symptoms like fever, nausea, vomiting, and seizures. In the pediatric population, approximately 30-50% of patients experience optic neuritis during their first NMOSD episode. While NMOSD typically targets the CNS, cases of NMO with

inflammatory pulmonary manifestations have been reported². IMT symptoms are nonspecific, varying with location, such as cough and chest pain for pulmonary IMTs and gastrointestinal symptoms for abdominal IMTs⁵. In this patient's case, symptoms primarily aligned with NMOSD, with finger clubbing being the sole pulmonary symptom. Given the correlation between NMOSD and other autoimmune inflammatory disorders, it's imperative to consider autoimmune or inflammatory pathologies during initial NMOSD diagnosis, particularly in younger patients.

NMOSD is a highly relapsing disorder and therefore long-term immunotherapy is required with rituximab, mycophenolate mofetil, and azathioprine being the most commonly used long term therapies in children. Acute episodes are treated with high dose prednisolone, plasmapheresis, or IVIG¹. IMTs are typically managed through complete mass resection with negative margins.

In conclusion, IMT of the lung and neuromyelitis optica spectrum disorders (NMOSD) are both rare inflammatory conditions with low incidence rates. This is the first reported case with pulmonary IMT along with NMOSD.

Resources

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